Techniques for Imaging Adult Congenital Heart Disease: CT Versus MRI

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Disclosures

• None
Introduction

• Pros/ cons of CT & MRI
• Common indications
• Technical pitfalls/ troubleshooting
Common Indications

• Aortopathy – aortic size, VTI, AR, LV mass
• TOF – RV size & function, TR, PR, PA stenosis
• TGA
  – Atrial switch: baffle obstruction/ leak, RV size and function
  – Arterial switch: supravalvular PS, aortic root dilation, coronaries, LGE
• Fontan – systemic ventricular failure, TE, obstruction of Fontan pathways
  – compression of pulmonary veins, etc.
Benefits of MRI/CT

MRI
• Volumetry – RV Vol/ EF
• Regurgitant Fractions
• Qp:Qs
• Temporal resolution

CT
• Coronary anomalies
• AP collaterals
• Breath-holding/ Sedation
• PPM/ AICDs /coils /clips

Extracardiac vasculature
Cost yield ratio
Bicuspid Aortic Valve

- AD inheritance/ spontaneous
- Stenosis, regurgitation, IE
- Aortic root and asc aortic dilatation – dissection
- Associated coarctation, VSD, PDA
- 25% need aortic surgery <25 yr/ 50% no significant disease
- CMR necessary to evaluate the entire aortic root from annulus to the arch
Aortic root/ Asc Aortic size/ LV Mass
BAV and Coarctation
Aortic Regurgitation

Forward flow: 116 mL
Ao SV = 84 mL
Regurgitation fraction 27%
LV SV = 152 mL
Aortic Regurgitation Fraction

• Aortic Regurgitation Fraction:
  \[
  \frac{140 - 90}{140} = 36\%
  \]

FF = 140 mL

Systemic SV: 53 + 37 = 90 mL
Marfan’s/Loey’s Dietz Syndrome

- Inherited disorders of connective tissue structure and function
- 1/3-5000
- Similar genotype
- FBN1/2 / TGFBR1/2
- Aortic dilatation and dissection
Marfan's Syndrome

- Skeletal, ocular findings
- Affects aorta, myocardium, valves and pulmonary arteries
- Mean age at first surgery 39 years
- Mean age at first dissection 39 years
- Surgical repair when external diameter of root/AA >5cm

Loey's Dietz Syndrome

- Hypertelorism, split uvula
- Widespread systemic aneurysms
- Mean age at first surgery 35 years
- Mean age at first dissection 38 yrs
- Surgical repair when external diameter of root/AA >4.4-4.6 cm
Arterial tortuosity and outcomes in Marfan syndrome and Loeys-Dietz syndrome

Increased Vertebral Artery Tortuosity Index Is Associated With Adverse Outcomes in Children and Young Adults With Connective Tissue Disorders

Shaine A. Morris, MD; Darren B. Orbach, MD, PhD; Tal Geva, MD; Michael N. Singh, MD; Kimberlee Gauvreau, ScD; Ronald V. Lacro, MD

Circulation 2011
Status at age 30 by VTI

- No surgery
- Prophylactic surgery
- Surgery for Dissection
- Dissection after surgery

<table>
<thead>
<tr>
<th>VTI</th>
<th>&lt; 20</th>
<th>20-39</th>
<th>&gt;=40</th>
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<tr>
<td>0%</td>
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<td>10%</td>
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<td>100%</td>
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- VTI: Volume of Tissue Involvement
CMRI Post Repair of TOF

- Gold standard for ventricular volumetry
- Every 2-4 years
- Therapeutic decisions: PV replacement (lasts 15-20 yrs), stented valve in conduit, AICD
- Timing of valve replacement: symptoms, progressive RV dilatation, progressive TR

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<thead>
<tr>
<th>Indications</th>
<th>Supporting References</th>
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<tr>
<td>1. Asymptomatic patients with ≥2 of the following criteria:</td>
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<tr>
<td>a. RV end-diastolic volume index &gt;150 mL/m² or z score &gt; 4.</td>
<td>10, 12</td>
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<td>In patients whose body surface area falls outside published normal data:</td>
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<tr>
<td>RV/LV end-diastolic volume ratio &gt; 2</td>
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<td>b. RV end-systolic volume index &gt;80 mL/m²</td>
<td>11, 13</td>
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<td>c. RV ejection fraction &lt;47%</td>
<td>11, 15, 16</td>
</tr>
<tr>
<td>d. LV ejection fraction &lt;55%</td>
<td>11, 15, 16</td>
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<tr>
<td>e. Large RVOT aneurysm</td>
<td>17, 18</td>
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<tr>
<td>f. QRS duration &gt;160 ms</td>
<td>11</td>
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<td>g. Sustained tachyarrhythmia related to right-sided heart volume load</td>
<td>6</td>
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<tr>
<td>h. Other hemodynamically significant abnormalities:</td>
<td></td>
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<tr>
<td>• RVOT obstruction with RV systolic pressure ≥0.7 systemic</td>
<td>19</td>
</tr>
<tr>
<td>• Severe branch pulmonary artery stenosis (&lt;30% flow to affected lung)</td>
<td>19</td>
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<tr>
<td>• Greater than or equal to moderate tricuspid regurgitation</td>
<td>19</td>
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RV Volumetry

RVEDV >160-170ml/m^2 – failure to recover post PVR
RV Diastolic Dysfunction
Tricuspid Regurgitation
Pulmonary Regurgitation
VSD Patch Leak
Branch PAs
Transposition of the Great Arteries

Anatomic correction (arterial switch)

• TGA with IVS (Jatene)
• TGA with VSD (Rastelli)

Physiologic correction (atrial switch)

• Senning
• Mustard
TGA, status post atrial switch, MRA
Systemic venous baffle obstruction

Cath. courtesy Dr. Nihill, Cardiology, Texas Children’s Hospital
Pulmonary venous baffle obstruction
Baffle Leak

- Patch leak may be seen on any view. Look for subtle dephasing
- Confirm using orthogonal view along jet core
- Free breathing flow velocity mapping to calculate $Q_p:Q_s$ ratio
Branch Pulmonary Artery Stenosis after ASO
Fontan - Complications

- Systemic ventricular dysfunction
- Thromboembolism
- Dilatation of the systemic venous atrium
- Obstruction of the Fontan pathways
- Pulmonary artery stenosis

- Compression of the pulmonary veins
- AV valve regurgitation
- Arrhythmias
- Protein-losing enteropathy
- Cyanosis
Baffle leak after Fontan
Failure of the Systemic Ventricle
Compression of pulmonary vein by dilated RA in AP Fontan
Thromboembolism in Fontan
Two techniques – dual injection/single injection delayed
Summary

• CT and CMRI – surveillance, targeted questions
• MRI gold standard for ventricular volumetry
• Surveillance of grafts, conduits and baffles
• Determination of timing of surgical intervention
References


Thank you!